

PERIPARTUM CARDIOVASCULAR DISEASE MINI-FOCUS ISSUE

ADVANCED

CASE REPORT: CLINICAL CASE

Successful TPV Implantation in a Pregnant Patient With Right Ventricle to Pulmonary Artery Conduit Obstruction



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ABSTRACT

A patient with repaired double outlet right ventricle presented during early gestation with heart failure symptoms due to severe right ventricle-pulmonary artery conduit stenosis and insufficiency. In the first trimester, she underwent transcatheter therapy with Melody pulmonary valve implantation with excellent hemodynamic results and completed pregnancy without significant maternal complications. (**Level of Difficulty: Advanced.**)

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In women with congenital heart disease (CHD), hemodynamic changes of pregnancy can threaten maternal and fetal life (1,2). Maternal cardiac output rises by 30% to 50% above baseline during pregnancy, thus increasing pressure overload from preexisting valve stenosis, leading to ventricular strain, heart failure, and arrhythmias. Mechanical relief of pressure overload due to valve stenosis should improve maternal outcomes. Most studies of valvular disease in pregnancy involve acquired aortic

or mitral valve disease, rather than CHD, with little research on pulmonary valve replacement (PVR) in pregnancy.

HISTORY OF PRESENTATION

A 20-year-old primigravida with repaired double outlet right ventricle presented at 6 weeks' gestation with dichorionic twins. The patient had a body mass index of 42 kg/m² with New York Heart Association functional class II symptoms and loud systolic and diastolic murmurs on examination.

LEARNING OBJECTIVES

- To understand the implication of conduit stenosis and valvular insufficiency in pregnant patients.
- To be able to weigh the risks and benefits of percutaneous valve replacement as a viable option in pregnant women with complex congenital heart disease.

MEDICAL HISTORY

Prior surgeries included a Rastelli procedure at age 4 years, when an aortic valve homograft was used for the right ventricle-pulmonary artery conduit (RVPAC) and at 12 years of age, the RVPAC was replaced with a 22-mm Contegra (Medtronic, Inc., Minneapolis, Minnesota) conduit.

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Informed consent was obtained for this case.

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ABBREVIATIONS AND ACRONYMS

CHD = congenital heart disease

CPB = cardiopulmonary bypass

PVR = pulmonary valve
replacement

RV = right ventricle

RVPAC = right ventricle-
pulmonary artery conduit

TPV = transcatheter pulmonary
valve

DIFFERENTIAL DIAGNOSIS

The differential diagnoses included heart failure from RVPAC stenosis and/or valve regurgitation, cardiomyopathy, pulmonary hypertension, sleep apnea or obesity hypoventilation, chronic pulmonary emboli, pregnancy-induced high-output heart failure, or tachyarrhythmias. Noncardiac differential diagnoses of shortness of breath on exertion included interstitial lung disease, anemia, and exercise-induced asthma.

INVESTIGATIONS

Echocardiogram confirmed significant RVPAC dysfunction with both moderate stenosis and regurgitation and an estimated right ventricle (RV) systolic pressure of 55 mm Hg. Given these findings in early gestation and the high likelihood of worsening heart failure as pregnancy progressed, the decision was made to replace her conduit to prevent pregnancy loss or maternal mortality. She thus underwent transcatheter pulmonary valve (TPV) implantation.

MANAGEMENT

At 13 weeks' gestation the patient underwent cardiac catheterization under general anesthesia. Radiation was minimized by using a low-pulse fluoroscopy rate of 3 frames per second. Baseline RV pressure was 70/18 mm Hg with a 42 mm Hg gradient across the RVPAC. Although some conduit calcification was noted, RVPAC angioplasty was performed with 20 mm × 4 cm and 22 mm × 4 cm GOLD ATLAS PTA catheters (Bard Peripheral Vascular, Inc., Tempe, Arizona), noting a waist at the annulus level but no waist at maximal inflation. RV pressure was still 64/16 mm Hg with a significant gradient at the level of the valve, as seen on angiography during conduit measurements (Figure 1). Because of the conduit regurgitation and persistent RV pressure elevation, we proceeded to TPV implantation after negative coronary compression testing.

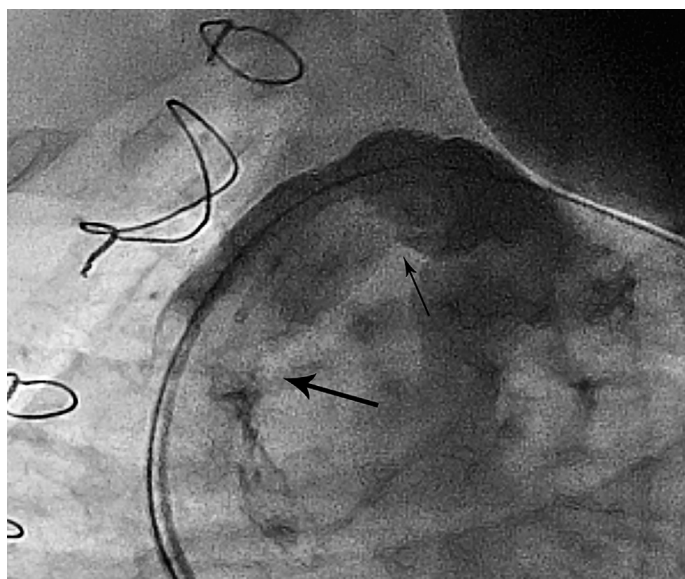
Pre-stenting was accomplished with consecutive delivery of 3 Palmaz bare-metal stents, each on a 22 mm × 4 cm Balloon-in-Balloon catheter (NuMed, Hopkinton, New York) through a 14-F Mullins trans-septal sheath. The first 2 stents, both Palmaz P4010 stents, were intended to build a longer landing zone, but a small gap remained between the two, so a third stent (Palmaz P3110) was implanted in telescopic fashion to bridge this gap. Subsequently, a 22-mm Melody TPV (Medtronic) was loaded onto a 22-mm Ensemble delivery system (Medtronic) and implanted inside the prestented landing zone. Final hemodynamics showed a significantly decreased RV pressure of 44/13 mm Hg, a total gradient of only 7 mm Hg across the RVPAC, and no residual valvular insufficiency (Figure 2). The patient tolerated the procedure well with no post-procedural complications.

The patient declined fetal echocardiography during pregnancy, but no gross anomalies were seen by routine fetal ultrasounds. At 23+4 weeks' gestation, the patient experienced intrauterine fetal demise of Baby A for unknown causes. At 30+6 weeks' gestation, she delivered a stillborn male and a healthy female infant without complication.

DISCUSSION

Until recently, valve replacement required open heart surgery with cardiopulmonary bypass (CPB). However, CPB during pregnancy is very high risk and is associated with a 30% fetal loss rate (3). Catheterization-based techniques are commonly used in rheumatic heart disease, and as a result,

FIGURE 1 Pre-Intervention Angiographic Image



Left anterior oblique view showing obstructed conduit inflow (thick arrow) and obstructed conduit outflow (thin arrow).

percutaneous mitral valvuloplasty is the intervention of choice for symptomatic mitral stenosis in pregnant patients (1). Sadler *et al.* (4) found a higher risk of fetal demise and preterm delivery in women with mechanical valves (59% and 57%, respectively) than in women with biologic valves (7% and 0%, respectively). Batra *et al.* (5) also reported a higher risk of fetal loss in women with mechanical valves than biologic valves (61% vs. 15%, respectively). Neither study included women with prior PVR.

In the modern surgical era, it is exceedingly rare for a patient to undergo mechanical PVR; nearly all performed today are with bioprosthetic valves. Since the Melody TPV was first developed by Bonhoeffer *et al.* (6) in 2000, TPV implantation has become a less invasive option than surgery in appropriate patients. Thus, Melody valve implantation allows insertion of a competent valve in a pregnant patient without the known risks of CPB to the fetus.

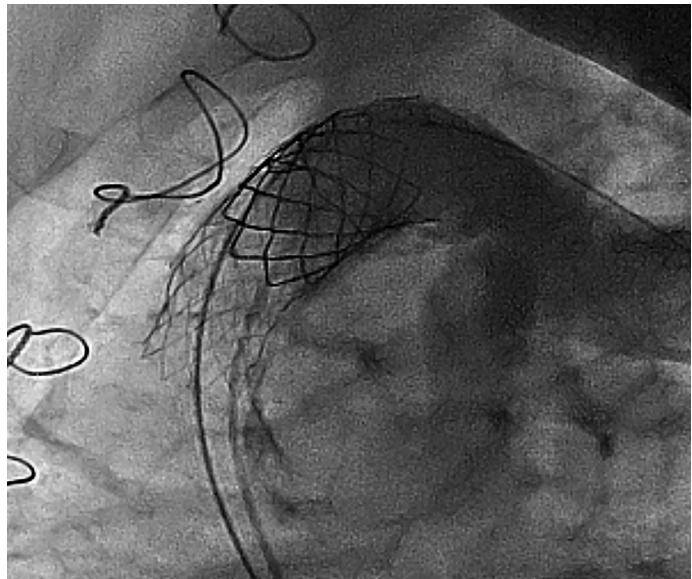
For our patient, conduit angioplasty alone was ineffective because of significant existing pulmonary regurgitation, which was no better or even worse after balloon dilation alone. Moreover, angioplasty did not effectively reduce her RVPAC gradient, so proceeding with valve replacement was deemed to be the best option. Without the availability of transcatheter valve therapy, our patient would have been presented with the difficult decision of the following: 1) accepting a 30% risk of losing her fetus during open heart surgery; 2) continuing pregnancy without intervention but with increased maternal risks and the high risk of premature delivery; or 3) terminating the fetuses to reduce the risk of a poor maternal outcome.

To our knowledge, there is only one other published case of successful TPV implantation during pregnancy (7). Although it is always preferable to perform congenital heart interventions in the nonpregnant state, an unfortunately large number of patients are “lost to care” for a variety of reasons and do not have valve reintervention done in a timely fashion before pregnancy. We believe this report highlights the usefulness of percutaneous valve therapy as an approach that provides more options to pregnant women with CHD.

FOLLOW-UP

The patient has been followed for 3 years since delivery and her most recent echocardiogram 2 years after TPV implantation showed an excellent result

FIGURE 2 Post-Intervention Angiographic Image



Left anterior oblique view showing a widely patent right ventricular outflow tract and no regurgitation at the level of the valve.

with peak and mean gradients of 17 mm Hg and 9 mm Hg, respectively, across the RVPAC and only trace regurgitation.

CONCLUSIONS

TPV implantation of a Melody valve can be safely done during pregnancy with excellent hemodynamic and pregnancy outcomes, as described in this report. Because of a growing population of patients with CHD reaching reproductive age, as well as the epidemic of patients “lost to follow-up” who present in an already pregnant state with significant valve problems, we expect use of TPV implantation techniques to become more common and crucial in reducing maternal and fetal mortality in the future.

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